Letters to the Editor

Fibrotic involution of sural muscles secondary to lupus panniculitis

Sirs

Lupus panniculitis (LP) is an inflammatory disease of the subcutaneous adipose tissue that can sometimes develop in patients with systemic lupus erythematosus (SLE) or, more frequently, discoid lupus erythematosus (DLE). LP can also occur as a primary entity in patients without features of LE or other autoimmune diseases (1-5). The disease course is chronic and characterized by remissions and exacerbations.

Hydroxychloroquine, corticosteroids, azathioprine, cyclosporin, thalidomide, cyclophosphamide are the drugs used for the treatment of LP either alone or in combination. A high percentage of patients require prolonged treatment for several months or years. Disfigurement and disability are relatively common in patients with LP. In the patient we describe a severe relapsing panniculitis, affecting multiple regions, resulted in a bilateral fibrotic involution of the sural muscles.

A 28-year-old man was referred to our department in November 2001 for subcutaneous, painful infiltrations of the abdomen, right thigh, calf and breast with overlying erythema, associated with fever (38-39°C). These symptoms had started 2 months before and the patient had been hospitalized twice elsewhere.

Histological examination of an abdominal subcutaneous fat nodule, performed during a previous hospitalization, showed the coexistence of large necrotic areas and extensive inflammatory infiltrations with the presence of lymphocytes, hystiocytes and neutrophils. The small and medium-sized vessels showed a slight infiltration of the intimal layer. In semi-thin sections a diffuse intimal thickening of the blood vessels was present with mild granulocytic infiltration. On electron microscopy the suppurative and necrotic areas appeared to be made up of neutrophils and histiocytes with the cytoplasm full of lipid inclusions. The surrounding areas displayed abundant plasma cells and fibroblasts. The walls of the small vessels were infiltrated by lymphocytes and neutrophils.

Physical examination on admission to our department did not reveal signs of visceral involvement. No clinical features of SLE or DLE were present. Chest radiography and computed tomography of the abdomen showed no abnormalities.

Laboratory data revealed high values of ESR (110 mm/1h) and CRP (191 ng/dl); high serum levels of fibrinogen (744 mg/dl), mild leukocytosis (9700 mm^3) and a normal number of platelets (276.000 mm^3). Serum levels of CPK, LDH, ALT, AST, alpha 1 antitrypsine, IgG, IgA, IgM, C3 and C4 complement components were normal. Assay for ANA (IIF on HEp-2 cells) was positive (1:160); antibodies to dsDNA, ENA and phospholypids were absent.

Treatment with prednisone (50 mg/day), hydroxychloroquine (400 mg/day) and cyclosporin A (200 mg/day) was started, resulting in disappearance of the fever, a rapid improvement of the subcutaneous lesions and a return to normal inflammation marker values. The patient was placed on maintenance therapy with an unmodified dose of hydroxychloroquine and cyclosporin A, while prednisone was gradually tapered to 10 mg/day.

In March 2002 the patient was re-admitted to our department because of the reoccurrence of symptoms: he had subcutaneous infiltrations in his right breast and calf associated with fever. High values of ESR, CRP and fibrinogen were present. Flexibility and stretching movements of his right foot were very limited and it assumed a club-foot position. MR scan revealed a reduced trophism of the back of the right leg with the presence of strongly hypointense areas due to fibrosis (Fig. 1). The treatment was modified. Hydroxychloroquine was replaced by azathioprine (100 mg /day) and prednisone by methylprednisolone (32 mg/day). Fever and pain disappeared and serum levels of inflammation markers returned to normal values.

Six months later the patient was admitted once again due to the worsening of his symptoms. On this occasion the left foot also presented a club-foot position. Ultrasound examination of the left leg showed the presence of fibrotic involution of the sural muscles.

Disability and disfigurement are not rare in patients with LP as a consequence of painful cutaneous lesions, residual cutaneous scarring and subcutaneous atrophy. In our patient, who was affected with a chronic, severe relapsing LP, disability and disfigurement were the result of a bilateral fibrotic involution of the sural muscles due to the spreading of subcutaneous adipose tissue inflammation. To our knowledge this is the first report of a case of irreversible muscle impairment due to LP.

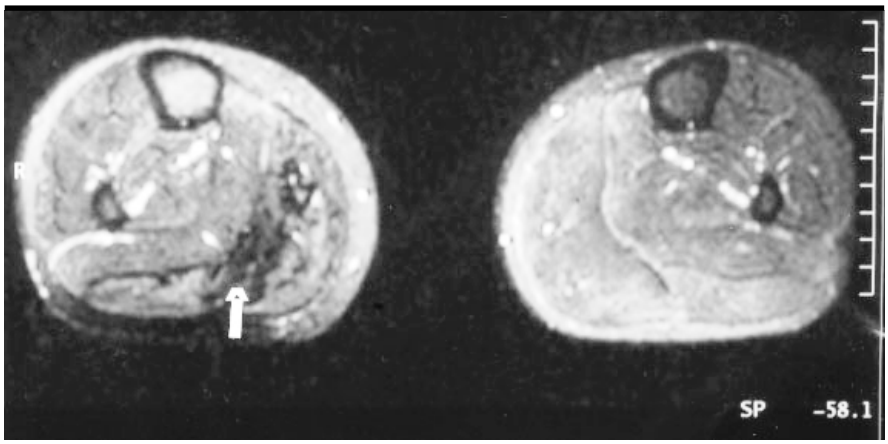

Fig. 1. MRI of the legs showing hypointense areas (**arrow**) on the back of the right leg due to fibrosis of the sural muscles.

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